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A rare case of solitary fibrous tumour of the sigmoid mesocolon: imaging features and review of literature

Ravikanth Balaji^a, Krishnankutty Ramachandran^a and Tara Somanathan^b

^aImageology Division, Regional Cancer Centre, Medical College PO, Trivandrum, Kerala, 695011, India;

^bPathology Division, Regional Cancer Centre, Medical College PO, Trivandrum, Kerala, 695011, India

Corresponding address: Ravikanth Balaji DNB, DMRD, Imageology Division, Regional Cancer Centre, Medical College PO, Trivandrum, Kerala, 695011, India.

Email: ravikanthbalaji@gmail.com

Date accepted for publication 7 September 2009

Abstract

Solitary fibrous tumours are rare spindle cell tumours that generally arise from the pleura but on rare occasions arise in other locations such as the head and neck region, stomach and retroperitoneum. Very few reports exist on the imaging findings of these tumours in extra pleural sites such as the mesentery. We report the computed tomography (CT) imaging findings of a rare case of solitary fibrous tumour in a 68-year-old man who presented with a slowly enlarging painless pelvic mass. CT scan showed a well-defined, multilobulated, highly vascular pelvic mass with dense calcifications and central hypodense areas arising from the mesentery.

Keywords: Solitary fibrous tumour; mesocolon; computed tomography.

Introduction

Solitary fibrous tumours (SFT) are rare mesenchymal neoplasms that typically originate in the visceral pleura^[1]. These tumours are also known to arise from extrapleural locations such as orbit, oral cavity, nasopharynx, larynx and the retroperitoneum but rarely from the mesentery^[2]. It is important to identify these tumours as they can pose a diagnostic dilemma for the clinician and radiologist alike. Moreover, there have been recent reports of some of these tumours exhibiting aggressive behaviour. A review of the literature yielded very few reports on the imaging of these tumours arising from the mesentery. We report a rare case of SFT presenting as a painless pelvic mass which at surgery was found to arise from the mesentery of the sigmoid colon.

Case report

A 68-year-old man presented with a large slowly progressive painless pelvic mass of 6 months duration with no

bladder or bowel symptoms. On physical examination, the mass was non-tender, firm and mobile. Ultrasound showed a large hetero-echoic mass in the pelvis superior to and abutting the bladder; Doppler interrogation revealed extensive vascularity. Plain computed tomography (CT) showed a large, multilobulated, heterogeneous soft tissue mass with dense calcifications and central rounded hypodense areas measuring $16 \times 12 \times 7$ cm arising from the mesentery. Contrast CT was performed in three phases in view of the increased vascularity seen on Doppler. Intense enhancement was observed in the peripheral solid portion on the arterial phase with gradual filling in of contrast in the portal venous phase. However, the central regions did not enhance suggesting these areas could have undergone necrosis, myxoid or cystic degeneration (Fig. 1A,B). Wash out of contrast was observed in the third phase (delay of 1 min after the portal venous phase). The mass was closely abutting the dome of the bladder and the sigmoid colon (Fig. 2). Anteriorly the mass reached up to the abdominal wall and posteriorly was in close contact with right ilio-psoas

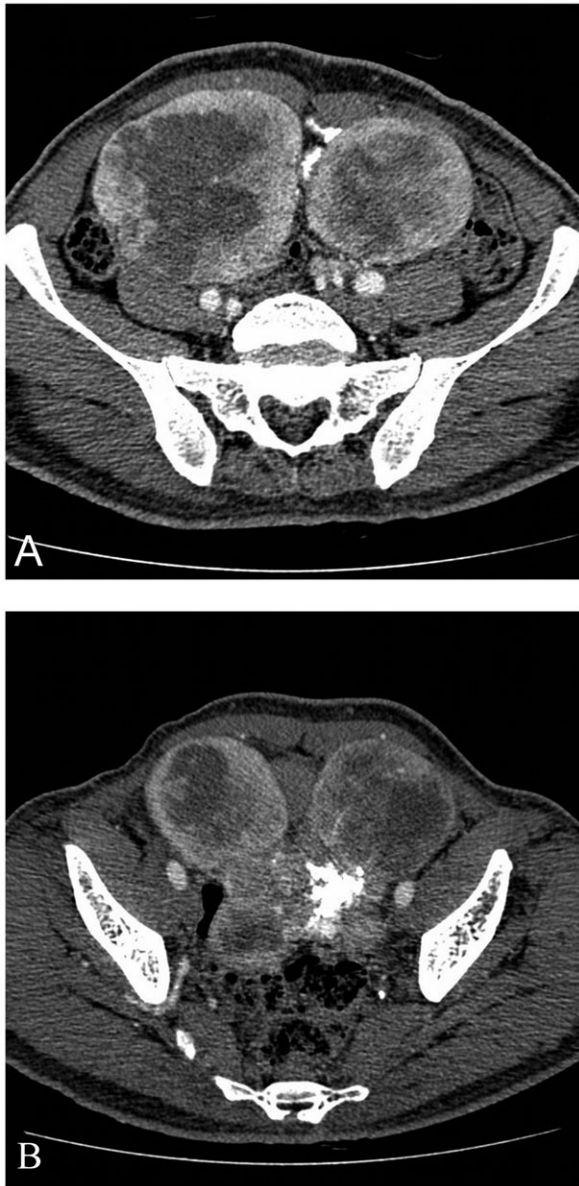


Figure 1 (A,B) Axial contrast CT images show a lobulated, enhancing, soft tissue mass with central hypoattenuating regions and dense calcifications.

muscle but with no infiltration. There was no peritoneal spread or vessel encasement. At surgery, a large multi-lobulated highly vascular tumour measuring 18×15 cm was seen arising from the lateral aspect of the sigmoid mesocolon. There was no attachment or infiltration to adjacent structures and the mass was resected totally. Histologic examination revealed solid areas with cystic spaces and calcifications with numerous small- and medium-sized branching stag horn vessels with hyalinized walls. Histopathology demonstrated a disorganized network of spindle cells in a background of collagen. Dilated vascular channels were seen interspersed with the spindle cells (Fig. 3). On immunohistochemical staining, the cells were positive for CD34



Figure 2 Coronal and sagittal contrast-enhanced CT images show the anatomic relationship of the mass with the bladder and rectum.

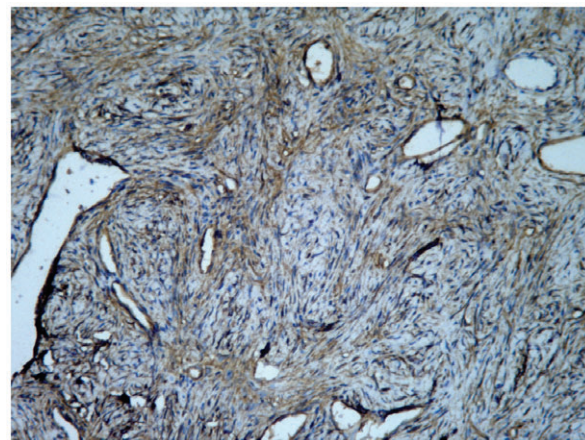


Figure 3 Photomicrograph showing spindle cells and endothelial cells lining the blood vessels with CD34 positivity (immunohistochemical staining, $\times 100$).

(transmembrane glycoprotein) and negative for smooth muscle actin and C-kit (CD117) confirming the diagnosis of a solitary fibrous tumour.

Discussion

SFTs are rare spindle cell neoplasms that typically arise from the pleura but have been reported in extra pleural locations such as the stomach, retroperitoneum, and head and neck regions^[2]. Patients usually present with slow-growing painless masses that manifest only when the tumours become huge. Rarely systemic symptoms such as hypoglycaemia, arthralgia and clubbing may be present. In a study of SFTs arising from the pelvis in males, the prostate was the most frequent organ of origin. Most SFTs are benign and have a favourable prognosis although there are reports of malignancy in

13–23% of cases^{1,3}. High cellularity, increased mitotic activity (more than 4 mitoses per 10 high-power fields), pleomorphism and haemorrhage are the factors favouring malignancy¹. SFTs occurring in extrapleural locations are mostly benign and are cured by surgical resection¹.

Benign SFTs of the peritoneum or mesentery have a favourable prognosis and do not recur after surgery¹.

On multi-slice CT imaging SFTs appear as heterogeneously enhancing multilobulated soft tissue masses with areas of dense or scattered calcification and central tubular or rounded hypoattenuating areas representing necrosis, myxoid or cystic degeneration. Enhancement is dependent on the cellularity and is most conspicuous in the arterial and early portal venous phases with wash out of contrast in the delayed phase. However, if fibrous elements predominate the contrast enhancement becomes marked in the delayed phases as well. On magnetic resonance imaging the tumours have a heterogeneous appearance on both T1- and T2-weighted images with the solid areas appearing iso- to hypointense to skeletal muscle on all sequences and the cystic areas appearing hyperintense on T2-weighted images. The contrast enhancement is similar to that seen on CT imaging. These tumours usually do not infiltrate but tend to displace and compress adjacent structures.

The differential diagnosis includes benign tumours involving the mesentery such as desmoid, inflammatory pseudo tumour, mesenteric fibromatosis and leiomyomas⁴. Gastrointestinal stromal tumour may be differentiated by the presence of extensive foci of haemorrhage and necrosis. Malignant lesions of the mesentery that have to be differentiated include soft tissue sarcomas, leiomyosarcomas, lymphoma and metastases. The presence of rounded necrotic areas, calcifications and the enhancement pattern in the arterial, portal venous and delayed phases aid in differentiating from other neoplasms arising from the mesentery.

Grossly, SFTs are well encapsulated and show a wide range of morphologic features depending on the degree of cellularity and vascularization and the fibrotic content. Histologically, these tumours show spindle cells arranged

in a haphazard manner with interspersed vascular channels in a collagenous background. Nuclear atypia or mitoses are not usually observed. Immunohistochemical studies are essential to differentiate from spindle cell tumours. The tumour cells are strongly positive for immunohistochemical staining with CD34 (transmembrane glycoprotein) and CD117 and negative for endothelial (CD 31), nerve sheath (S-100) and myogenic (smooth muscle origin) markers^{5,6}.

SFTs are benign and can be surgically resected. They do not usually recur or metastasize. However, if the pathology reveals malignant features close follow-up is needed.

In conclusion, we have reported the imaging features of a rare SFT arising from the mesentery. The tumours are typically vascular and enhance intensely with contrast and show necrosis or cystic degeneration and calcification. Radiologists must be aware of this entity when evaluating unusually large enhancing masses in the pelvis and SFTs should be considered in the differential diagnosis of large pelvic tumours.

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